

A Case of Rhizomelic Spondylosis

Service of Dr. Charles K. Mills

Reported by Dr. Ralph Pemberton, Resident Physician



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The patient's father was a Russian, and died at 49 years of age, after an illness of one month in bed, with swelling of the legs.

The patient states that he has been told that his father was subject to rheumatism. His mother is living and well at 50 years of age, and two sisters, aged respectively, 35 and 37, and one brother of 25 years of age, are also in good health. There has been no other death in the family, and apparently no family diseases.

The patient was born in Russia; denies all diseases of childhood except measles, and states that he has had no venereal disease or illness of any other kind beyond the present one, and had never been in a hospital until admitted for his present complaint.

He used liquor very moderately and tobacco to slight excess.

Nine years ago he was married, and has three healthy children, his wife being a healthy woman, having had no miscarriages as far as he knows. No children have died.

He came to this country when 16 years of age, and for about four years worked steadily at cap-making, bending over his work all day long and getting no exercise; his duties consisting in feeding the various pieces of cloth to the machine, which sewed them together.

He next took up huckstering, at which occupation he worked for one year, and did a great deal of walking, pushing a cart around the city or standing in one place with his wares, always out in the open, but never exposed, he says, to very stormy weather.

In the course of six months he began to feel below par, though just how is hard to determine beyond the fact that he had pain in his right leg from the hip to the knee. In a few months more his left leg became similarly affected, and the pains in both increased in severity, though not sufficiently to cause him to cease work, and it cannot be ascertained that any other parts of his body were then involved. Shortly, however, after a total of one years of out-of-door work and exercise, he was admitted to the Jewish Hospital, where, with one or two brief intermissions, he has been for nearly three years. While there he says he developed stiffness of the shoulders, both hips, neck, back and knees, though he does not remember the order of involvement, or even which was first. He was in bed for most of his stay at the hospital, but for the last few months had spend the days in a wheel chair.

Examination.—A moderately emaciated male of about 35 years of age, with light-brown hair and intelligent expression. He wrinkles brow in all directions, draws mouth to both sides, shows teeth evenly and whistles normally. Patient stands without support, sways slightly with the eyes closed and slightly more with the eyes closed very tightly.

As he stands the legs are held stiffly, the trunk is inclined forward from the hips and the upper portion of the trunk with the head is thrown directly forward to an angle of about 120 degrees with the rest of the body. The neck is almost perfectly rigid, and the man's body can be raised by raising the head with a hand under the occiput, the only possible motions being a rotation of the head of about 15 degrees to the right and about 45 degrees to the left, and a slight depression and elevation of the same.

The eyes look toward the floor except that, when he is talking to some one, they are rotated upward to counteract the downward inclination of his head, and enable him to look anteriorly on a level with his own head. The jaws can be opened fully wide and can be closed tightly.

The trunk cannot be straightened, and he cannot walk without the assistance of crutches, but with them he pushes his feet forward one after the other, bending the knees but slightly, and apparently not moving the legs at the hip joint. After a few steps of this kind he swings both the limbs forward together in a pendulum-sort of gait.

In sitting there is no movement at the hip joint whatever, the body resting on the chair with the tuber ischii almost on the edge of the seat and the shoulders against the uppermost part of the back of the chair, other portions of the body not touching it.

In rising the patient lifts himself by the arms, making no attempts to bend the knees as a normal individual would, but rather keeping himself from slipping back into the chair by fixing the heels against the floor. The eyes show moderately dilated, but equal pupils, which react promptly to light and accommodation, and the extra-ocular movements are good in all directions. The tongue is easily and evenly protruded and of normal aspect. The pulse is regular and of good volume.

The chest presents great deformity, giving the appearance of being sunken in at the sides, and long and narrow, while the clavicles extend almost directly backward instead of laterally, and slightly backward from the sternoclavicular articulation. The shoulder joints are unduly thrust forward, the spaces between the heads of the humeri and the clavicles very sunken, and the shoulders, as a whole, are extremely narrow, the most anterior parts of the humeri being but 25 cm. apart. Posteriorly there is bulging about the angle of the ribs on both sides, particularly the left, throughout the whole length of the thorax, making the posterior aspect flatter than normal and broad at the base.

From the back the sides of the chest run abruptly forward and inward, making an angle of about 40 degrees with the plane of the back, so that a cross section would give a figure broad of base, the sides slanting at an angle of about 40 degrees to the apex, which is about one-half the length of the base line.

The expansion of the chest seems about equal and slight on both sides. The apex beat is rather diffuse and easily visible in the fourth interspace, and one inch outside of mammary line.

The apex beat is forceful and diffuse, but without thrill. Cardiac dullness is easily elicited, extending from the third rib and left edge of sternum to about one inch outside of the nipple line. The first apical sound is of very fair timbre and the second valvular sound apparently normal. The lungs give a good note anteriorly to percussion, and seem clear on auscultation. The breath sounds are easily audible and rather harsh.

The abdomen is slightly, but not unduly protuberant, bulging moderately in the flanks. It is soft except when he lies on his back, and his head is unsupported, when the abdominal muscles assist apparently in maintaining his position, though without conscious effort on his part. As soon as the head is well supported they relax.

Both arms are somewhat wasted, the right considerably so, and the bellies of the biceps muscles, particularly the right, stand out prominently on either side. The right arm cannot be circumducted at the shoulder, and attempts at motion, passive or active, beyond the angle of 50 degrees with the axis of the body, either laterally or antero-posteriorly, meet with pain and firm resistance, both apparently in the shoulder articulation. The right arm cannot be fully flexed, the tips of his fingers being 5 cm. from the point of the shoulder when flexion is greatest. The motions of the left arm are fairly good in all directions, except directly upward, and he can touch the point of his left shoulder easily with the tips of his fingers. There is distinct wasting of the muscles of the right shoulder joint, particularly the supra- and infra-spinati, the deltoid, the biceps, trapezius and the latissimus dorsi. There are no fibrillary movements, but at times there seem to be fascicular tremors in the muscles of the right upper arm and in the infra-spinatus and pectoral regions.

The grasp of the two hands is very good, while the movements of the forearm and hand can be well performed; but muscular power in the upper extremities is much below normal—that of the right arm being much less than that of its fellow. There is no motion at all at the hip joint, and no atrophy of the lower extremities. The knees can be bent until the legs make an angle of about 45 degrees with the thighs, but effort to proceed further causes pain. Motion of the feet, ankles, toes, hands, wrists and fingers seems good, and there appears to be no wasting of the muscles connected with them. The reflexes of the upper extremities are apparently normal, and the patellar reflexes are present, but the excursion of the limbs is not very great because of the rigidity of the knee joints. Plantar flexion of the great toe results from all forms of stimulation to that end, and there is no ankle clonus. Achilles jerk is present and sensation is apparently normal everywhere in all its forms. Examination of his urine yields negative results. Since admission to the hospital the patient has had no rise in temperature. His bowels have been regular, and he has eaten and slept well.

FIG. I.



FIG. 2.



FIG. 3.



